POSTER SESSION

1171 Pediatric Electrophysiology: Acquired Heart Disease in Children

Tuesday, March 09, 2004, 3:00 p.m.-5:00 p.m.
Morial Convention Center, Hall G
Presentation Hour: 3:00 p.m.-4:00 p.m.

1171-199 Neurological Outcome of Fetal Arrhythmias Complicated by Hydrops

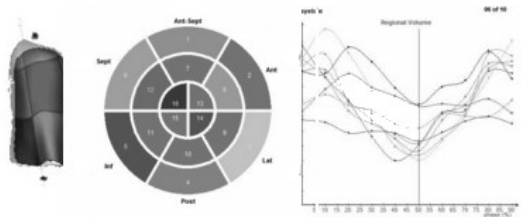
Johannes M. Breur, Martijn A. Oudijk, Rob H. Gooskens, Philip Stoutenbeek, Linda S. de Vries, Gerard H. Visser, Erik J. Meijboom, University Medical Center Utrecht, Utrecht, The Netherlands, University Hospital of Vaud, Lausanne, Switzerland

Objectives:
Fetal arrhythmias are associated with congestive heart failure and development of fetal hydrops, which may result in neurological morbidity and mortality. Limited data exists on the long-term outcome of hydropic fetuses.
Methods:
A retrospective study on cognitive and neurological functioning of 16 infants, aged 0,5 to 12 years, who experienced fetal arrhythmias complicated by hydrops.
Results:
Seven fetuses had supraventricular tachycardia, 3 had atrial flutter, 1 had ventricular tachycardia and 5 had congenital complete atrioventricular block (CCAVB). Mean GA at birth was 35 weeks and 5 days. Nine fetuses with tachycardia converted to sinus rhythm in a mean time of 7,9 days; resolution of hydrops was achieved in 6 of these patients in a mean time of 7,4 days.
Neonatal cranial ultrasound was normal in 7 infants and all but one of these were normal at follow-up: one infant showed a focal thalamic infarction, first seen by the end of the first week, and developed multiple cerebral lesions as a result of a malignant LQTS and died at the age of 2 years. Five infants had flaring on neonatal cranial ultrasound, one associated with a subependymal pseudocyst, and one with calcifications due to a congenital CMV infection. Two of these infants were normal at follow-up, one died two days after birth as a result of withdrawal of therapy, and one infant showed mild global delay. One infant showed evidence of a parenchymal haemorrhage of antenatal onset, presenting as a unilateral porencephalic cyst. He developed a mild hemiplegia with normal cognition. Three infants with CCAVB without cranial ultrasounds were normal at follow-up.
Conclusions:
Fetal arrhythmias complicated by hydrops are thought to predispose the unborn child to neurological damage. However, in this series 13 out of 16 infants were neurologically normal. Prognosis seems particularly good in case of successful treatment of tachycardia, delivery at term, and in case of CCAVB. Initiation of therapy should not be withheld or delayed on the assumption of poor neurological outcome.

1171-200 Real-Time Three-Dimensional Echocardiographic Evaluation of Global and Regional Left Ventricular Systolic Function in Young Patients Paced for Congenital Complete Heart Block

Swati Garekar, Kavitha Chintala, Peter Karpawich, Michael Pettersen, Childrens Hospital of Michigan, Detroit, MI

BACKGROUND: Pacing induces an abnormal activation sequence resulting in left ventricular (LV) asynchrony and sub-optimal contractility. This is important in young patients (pts) with congenital complete heart block (CCHB), who need long term pacing. Objective methods to assess altered global and regional LV function are limited. Real time Three-Dimensional echocardiography (RT3DE) images the entire ventricle without the need for geometric assumptions. We assessed the feasibility of RT3DE to quantify LV systolic function in paced CCHB pts.
METHODS: Volumetric data sets were obtained by RT3DE over 7 cardiac cycles with a matrix phased array transducer from the apex. Offline analysis utilized RT3DE software (TOMTEC). Using semi-automated border detection, a dynamic 16-segment wire-frame model of the LV cavity was constructed. Instantaneous segmental volume and ejection fraction curves were generated.
RESULTS: RT3DE was performed in 5 pts (median age 15y). Each data set was acquired over 7-10 seconds. 80% of data sets were suitable for analysis. Mean offline analysis time was 20 minutes per pt. Segmental volumes and ejection fractions were examined at end systolic and mid systolic phases and were used to assess synchrony in each pt. The intra-observer reliability was high (mean correlation coefficient 0.86).
CONCLUSIONS: This initial study demonstrates that RT3DE can be effectively used to evaluate LV segmental contraction in young paced patients with CCHB.



1171-201 Lone Atrial Fibrillation in Adolescent Patients

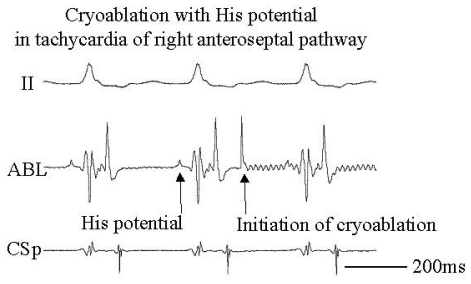
Nandini Madan, Victoria L. Vetter, Tammy Wieand, Ronn E. Tanel, Larry A. Rhodes, Childrens Hospital of Philadelphia, Philadelphia, PA

Background Atrial fibrillation (AF) is uncommon in children without heart disease. There are no natural history studies of this arrhythmia. **Methods** Patients less than 20 years of age, presenting with the first episode of AF, between January 1990 and April 2003 were included. Exclusion criteria were structural heart disease, presence of preexcitation and other atrial arrhythmias. Clinical, echocardiographic and electrocardiographic data was retrospectively reviewed. **Results** 15 patients (100% male) presented at a mean 15.9±1.7 years. 88.2% were symptomatic at presentation (8 palpitations,7 presyncope or syncope, 2 dyspnea). Systolic hypertension was present in 3 patients, 2 had hypotension. Associated diagnosis were leukemia (2), renal failure (2), connective tissue disorder (1), cardiac trauma (1), pulmonary hemorrhage (1), seizure disorder (1), asthma (1) and long QT syndrome (1). Ventricular rate at presentation was 125±14.5 bpm. Conversion to sinus rhythm was achieved electrically (5), pharmacologically (4) and spontaneously (6). In sinus rhythm the maximum P wave duration was 117 ±20(normal 96±11 msec) and P wave dispersion was 42.7± 21 msec (normal 38±10msec). On EKG 3 patients had incomplete right bundle branch block and 3 had left ventricular hypertrophy. The shortening fraction was 32.7± 14%. 1 patient had clot in the left atrium. Patients were followed for 33.2± 26.9 months. Chronic therapy consisted of digoxin (11) for 17±22 (1 to 61) months and beta blockers (2). 4 patients received no therapy **Conclusions**-Lone AF in adolescents is predominantly a male disease.In medium-term follow up symptomatic recurrences are rare so antiarrhythmic therapy is not indicated. Abnormalities in non-invasive markers of intraatrial conduction (P wave dispersion)may represent changes in substrate predisposing to recurrences in prolonged follow-up.

1171-202 Cryoablation for Septal Tachycardia Substrates in Pediatric Patients

Aya Miyazaki, Andrew D. Blaufox, David Fairbrother, J. Philip Saul, Medical University of South Carolina, Charleston, SC

Background: Catheter based cryoablation (CA) may be useful for ablation of septal tachycardias, including AV nodal reentry tachycardia (AVNRT), AV reciprocating tachycardia (AVRT) and junctional ectopic tachycardia (JET). However, minimal data on its safety and effectiveness exist in pediatric pts.
Methods: Ten pediatric pts (age: median = 13 yrs, 7-15; wt: 49 kg, 27-60) with septal tachycardias underwent CA.Four had AVNRT, 5 AVRT (n=4, right anteroseptal (RAS); n=1, right midseptal) and 1 JET. CA was performed via the IVC in 8 pts, SVC in 1, and the aorta in the JET pt. Applications were considered cryomaps (CM) if they were < 120 s or if the min tip temperature was > -35^{o}C. Other lesions were considered CAs.
Results: 69 CMs (3/pt, 1-21) and 23 CAs (1.5/pt, 1-4) were performed, for a total cryo time of 597 s (345-2449). Procedural success was achieved in 7 of 10 pts (3/4 AVNRT, 3/5 AVRT, 1/1 JET). Cryo was performed in tachycardia 32/92 times, resulting in 3 successes and 4 transient successes. In 2 pts (1 RAS pathway, 1 JET), success was achieved in the presence of a His potential on the ablation catheter (Figure). For pts with AVRT, permanent success occurred earlier after reaching –25^{o}C than transient success (0.9±2.1 vs 24.8±25.5 s, p=NS). There were no complications.
Conclusion: CA can be used safely and effectively to ablate septal tachycardias in pediatric pts, even in very close proximity to the His bundle and during tachycardia. Earlier success after tissue freeze may be predictive of a sustained effect.



1171-203 A Study of Left Ventricular Function in Children With Persistent Coronary Arterial Aneurysm Long After Acute Kawasaki Disease by Dobutamine Stress Echocardiography

Ling Hui, Maurice P. Leung, Adolphus KT Chau, Yiu-Fai Cheung, The University of Hong Kong, Hong Kong, Hong Kong

Background: Persistence of coronary arterial aneurysm (CAA) in patients after acute Kawasaki disease (KD) may result in myocardial ischemia, infarction and even sudden death. Dobutamine stress (DS) echocardiography was used to assess left ventricular (LV) function [wall motion abnormalities (WMA) and LV contractility] in this group of patients.
Methods: Twenty-two patients (11.3±4.3 years, 17 males) who had KD with CAA persisting for 8.49 years were studied. The mean aneurysmal diameters were 4.73±1.95 mm (ranged 2.8 to 8.8) at the time of study. For the patients without WMA, the LV contractility

were further assessed by fractional shortening (FS), ejection fraction (EF), stress-velocity index [rate-corrected velocity of circumferential fibre shortening (Vcfc) *vs* wall stress (WS)], both at rest and under DS (5μ g/kg/min). The results were compared to those of 20 normal subjects aged 10.4±4.5 years (p=0.52). All patients had their perfusion examined by thallium scan. WMA identified echographically were compared with perfusion defects.

Results: The patients were divided into 2 subsets: those with (n=13) and without WMA (n=9). For those with WMA (59%), ten had impaired myocardial perfusion. The areas of WMA correlated well with perfusion defects identified by thallium scan (r= 0.86). For those without WMA (41%), the FS (resting: 0.36±0.05 *vs* 0.41±0.05, p=0.033; stressed: 0.45±0.06 *vs* 0.54±0.04, p<0.001), EF (resting: 0.74±0.06 *vs* 0.80±0.06, p=0.022; stressed: 0.83±0.06 *vs* 0.90±0.03, p<0.001), Vcfc (resting: 1.13±0.19 *vs* 1.23±0.21, p=0.047; stressed: 1.41±0.18 *vs* 1.73±0.26, p<0.001) and WS (resting: 69.95±14.36 *vs* 72.67±11.65, p<0.001; stressed: 41.78±16.18 *vs* 46.2±15.98, p=0.048) were significantly different with the controls. The thallium scan revealed no myocardial perfusion defects, their LV contractility indices were significantly lower (p<0.05) than normal control at rest and under DS.

Conclusions: For KD patients with persistent CAA, LV contractility is abnormal in a significant proportion of patients, albeit asymptomatic. The presence of significant WMA in 59% of patients under DS suggests a need for long-term follow up of their LV function.

1171-204 Elevated hs-C-Reactive Protein Levels and Increased Arterial Stiffness in Children With a History of Kawasaki Disease

Yiu-fai Cheung, Marco H. Ho, Sidney C. Tam, Tak-cheung Yung, Adolphus K. Chau, The University of Hong Kong, Hong Kong, Hong Kong

Background - Increased arterial stiffness has been documented years after Kawasaki disease (KD). We hypothesized that low grade inflammation persists long-term after resolution of the acute phase and impacts on arterial stiffness in children with a history of KD. **Methods** - A cohort of 106 subjects was studied, which comprised 43 KD patients with coronary aneurysms (group I), 28 KD patients with normal coronary arteries (group II) and 35 healthy age-matched children (group III). We compared the systemic blood pressure, fasting serum cholesterol, high sensitivity-C-reactive protein (hs-CRP) level and carotid artery stiffness index among the three groups. We further interrogated the relationship between hs-CRP level and arterial stiffness and identified significant determinants of carotid arterial stiffness. **Results** - Serum hs-CRP level of group I subjects (median 0.39 mg/l, inter-quartile range 0.28-0.65 mg/l) was significantly greater than that of group II (median 0.24, inter-quartile range 0.17-0.29 mg/l, p<0.001) and group III subjects (median 0.25 mg/l, inter-quartile range 0.18-0.40 mg/l, p<0.001). Likewise, carotid artery stiffness index of group I subjects (5.07±1.11) was significantly greater than those of group II (4.27±0.83, p=0.002) and III subjects (4.24±0.86, p=0.001). For the entire cohort, carotid artery stiffness index correlated positively with log serum hs-CRP level (r=0.24, p=0.013). Using multiple linear regression analysis, age (standardized β=0.22, p=0.02), systolic blood pressure (standardized β=0.28, p=0.01), log serum hs-CRP level (standardized β=0.21, p=0.017) and patient grouping (standardized β=-0.36, p<0.001) were found to be independently associated with carotid artery stiffness. **Conclusion** - Our findings suggest ongoing low-grade inflammation late after resolution of the acute phase of KD in patients with coronary aneurysms. Furthermore, this low-grade inflammation may play a role in the increase of systemic arterial stiffness.

1171-205 Infantile Kawasaki Disease: High Incidence of Coronary Artery Aneurysms in Spite of Appropriate Intravenous Immunoglobulin Treatment

Teiji Akagi, Wakako Himeno, Masahiro Ishii, Hirohisa Kato, Toyojiro Matsuishi, Kurume University, Kurume, Japan

Background: Previous studies have demonstrated that Kawasaki disease (KD) in infants (< 3 months) is rare condition, however it usually takes severe and atypical clinical course with high incidence of coronary artery aneurysms. After the introduction of intravenous immunoglobulin (IVIG) treatment for the acute KD, the incidence of coronary lesions was significantly decreased, however little information was available about the clinical features in this patient population after the introduction of IVIG treatment. **Methods:** From January 1995 though December 2002, 750 patients with KD were experienced in our hospital, of which 25 infants (3.3%) suffered from KD <3 months of age. Their clinical feature was retrospectively analyzed. **Results:** Patients consisted of 15 boys and 10 girls. Median onset of disease was 72 (ranged from 37 to 86) days after birth. Patients were admitted on 2.8±1.3 days of illness. Only 11 infants were suspected of KD at the time of admission. Diagnosis was completed on 4.5±1.8 days of illness. The lacking items of diagnostic criteria were cervical lymphadenopathy (n=11), change of extremities (n=7), skin rash (n=1) and bilateral conjunctivitis (n=1). IVIG was indicated in 23 infants (2g/kg/day in 19, 400mg/kg/day for 5 days in 4) and initial treatment was started on 4.4±1.1 days of illness. Aspirin was given to all 25 patients. Initial IVIG treatment was failed in 9 infants, of which 7 infants were treated with intravenous methylprednisolone pulse therapy. Finally, 11 (44%) of 25 infants were complicated with coronary arterial lesions, including 8 with coronary aneurysms and 3 with transient dilatation. Non-cardiovascular complications, such as aseptic pyuria (n=7), aseptic meningitis (n=3), disseminated intravascular coagulation (n=2), hypotension or apnea during IVIG infusion (n=2), convulsion (n=1) or jaundice (n=1) were mostly found within 5 days of illness. **Conclusion:** In spite of the appropriate IVIG treatment was used in these infants, the incidence of coronary artery aneurysm was still high. Early diagnosis and other therapeutic strategies should be required for the improvement of cardiovascular outcome in these high-risk patients.

1171-206 Frequency of Infective Endocarditis Among Infants and Children With Staphylococcus Aureus Bacteremia

Anne Marie Valente, Rajiv Jain, Mark Scheurer, Vance Fowler, G. Ralph Corey, A. Resai Bengur, Stephen Sanders, Jennifer Li, Duke University Medical Center, Durham, NC

Background: The reported prevalence of infective endocarditis (IE) among patients with *Staphylococcus aureus* bacteremia (SAB) varies widely. In adults with SAB, the incidence of definite IE evaluated prospectively is 12.5%. The objective of this study was to determine prospectively the prevalence of IE among pediatric patients with SAB in a large tertiary care center.

Methods: Between July 1, 1998 and June 5, 2001, 66 children hospitalized at Duke University Medical Center developed SAB. Of these patients, informed consent was obtained and echocardiography was performed in fifty-one cases (77%). Demographic information, predisposing conditions, and blood culture results were collected. The patients were clinically evaluated for signs and symptoms of IE. Endocarditis was classified as definite, possible, or rejected based on modified Duke Criteria.

Results: Definite or possible IE was present in 10/51 (20%) children with SAB (6 definite, 4 possible). Most children (73%) developed bacteremia as a consequence of an infected intravascular device. Children with underlying congenital heart disease had a significantly higher prevalence of definite or possible IE, compared to those with structurally normal hearts (9 of 17; 53% vs. 1 of 34; 3%, p = 0.02). All of the patients with definite IE had multiple positive blood cultures with *S. aureus*. Mortality was higher among IE patients (4/10, 40%) than SAB patients without IE (9/41. 22%).

Conclusion: The prevalence of IE among children with SAB is 20%. IE due to *S. aureus* is frequently associated with congenital heart disease and multiple positive blood cultures. The mortality for children with SAB and definite or possible *S. aureus* IE is high.

ORAL CONTRIBUTIONS

857 Late Outcomes in Repaired Congenital Heart Disease

Tuesday, March 09, 2004, 4:00 p.m.-5:00 p.m.
Morial Convention Center, Room 271

4:00 p.m.

857-1 Neurodevelopmental Outcome After Repair of Total Anomalous Pulmonary Venous Connection

Paul M. Kirshbom, Thomas B. Flynn, Richard F. Ittenbach, Diane M. Hartman, Nancy B. Burnham, Ronn E. Tanel, Steven M. Paridon, Thomas L. Spray, Gil Wernovsky, J. William Gaynor, Childrens Hospital of Philadelphia, Philadelphia, PA

Background: Early survival after Total Anomalous Pulmonary Connection (TAPVC) repair has improved significantly. Little data are available concerning subsequent neurodevelopmental outcome.

Methods: School age survivors of TAPVC repair performed between 1/1983 and 12/1996 at our institution were eligible. Assessment included neurologic examination, cognitive function [Full Scale IQ (FS), Verbal (V), and Performance (P) IQ], fine motor skills (Grooved Pegboard) and visual-motor integration (VMI).

Results: Thirty children were tested at a median age of 11 yr (range 6-19). Median age at TAPVC repair was 16 days (range 1-141). TAPVC was supracardiac in 14, infracardiac in 12, cardiac in 3, and mixed in 1. Obstructed TAPVC occurred in 6 patients. Circulatory arrest was used for all repairs with a median duration of 35 min (range 17-66). Reoperation had been performed in 7 patients. At follow-up, microcephaly (head circumference ≤ 5%) was present in 28%. Neuromuscular examination was suspect or abnormal in 27%. Hyperactivity and/or attention deficits were present in 50%. Mean FSIQ (95.3 ± 18.5), and VIQ (98.6 ± 20.2) were not significantly different from population norms, both p > 0.1. Mean PIQ (92.3 ± 16.9) was significantly lower than the population norm, p=0.01. Fine motor skills (including fine motor speed, dexterity, VMI and hand-eye coordination) were significantly impaired compared to population norms (all p < 0.02). Younger gestational age and reoperation predicted lower FSIQ, VIQ, and PIQ, all p ≤ 0.05. Obstructed TAPVC also predicted a lower PIQ, p = 0.05. Age at surgery, length of stay at the initial surgery, and duration of circulatory arrest were not predictive of any adverse outcomes.

Conclusion: In school age survivors of infant TAPVC repair, there is a significant incidence of neurodevelopmental disabilities. Microcephaly is common. Fine motor skills and VMI are particularly concerning. There is also a frequent occurrence of hyperactivity and/or attention deficits.

4:15 p.m.

857-2 Long-Term Noninvasive Electrophysiologic Assessment After Repair of Total Anomalous Pulmonary Venous Connection

Ronn E. Tanel, Stephen M. Paridon, Paul M. Kirshbom, Diane M. Hartman, Nancy B. Burnham, Richard F. Ittenbach, Thomas L. Spray, J. William Gaynor, The Children's Hospital of Philadelphia, Philadelphia, PA

Background: Pediatric patients with a history of atrial surgery are at risk for the development of sinus node dysfunction (SND) and atrial arrhythmias. However, there has been no comprehensive, long-term, electrophysiologic (EP) study of patients who have undergone repair of total anomalous pulmonary venous connection (TAPVC). **Methods**: We evaluated school-age and adolescent survivors of isolated TAPVC repair from 1/83-12/96